



Uncontrolled hypertension in Fabry disease

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Abstract: Fabry disease is a x-linked lysosomal storage disease leading to early death related to renal, cardiac, and cerebrovascular disease. Therefore, proper diagnosis and therapy of elevated blood pressure may improve morbidity and mortality of these patients. However, the prevalence of uncontrolled hypertension in Fabry disease is unknown. We examined blood pressure of patients with Fabry disease using a large international database, the Fabry Outcome Survey (FOS). We defined uncontrolled hypertension as a systolic blood pressure (SBP) ≥ 130 , and/or a diastolic blood pressure (DBP) ≥ 80 mmHg (threshold for blood pressure control in renal disease, JNC7). We used the short MDRD-GFR formula for assessment of renal function, and we classified chronic kidney disease according to K/DOQI. Among 459 patients with Fabry disease, 306 had blood pressure readings entered in the database. Mean SBP was 124.6 ± 16.9 mmHg and mean DBP was 73.6 ± 11.7 mmHg (mean age: 38.4 ± 15.6 years, 142 females, 164 males). Forty-three percent of men and 28% of women showed uncontrolled hypertension. In 291 patients both, blood pressure readings and GFR estimates, were available. In patients with normal GFR (>90 ml/min/1.73m²) mean SBP was 119.5 ± 15.6 mmHg and mean DBP was 69.7 ± 11.1 mmHg (n=120). In patients with mild decreased GFR (60-89 ml/min/1.73m²) mean SBP was 126.7 ± 15.9 mmHg and mean DBP was 75.0 ± 11.0 mmHg (n=110). In patients with moderate decreased GFR (30-59 ml/min/1.73m²) mean SBP was 132.7 ± 20.8 mmHg and mean DBP was 79.0 ± 13.3 mmHg (n=41). In 70 patients blood pressure readings were available before start of enzyme replacement therapy (ERT) with agalsidase alfa (Replagal, TKT 5S Europe, 0.2 mg/kg bodyweight fortnightly i.v.), in 87 at 12 months and in 76 at 24 months of therapy. At baseline, at 12 and at 24 months of ERT, 39%, 30% and 42% of the patients presented with uncontrolled hypertension, respectively. Our study revealed a high prevalence of uncontrolled hypertension among patients with Fabry disease. Thus, there is need for improvement of blood pressure control in these patients. *Am J Hypertens* (2004) 17, 206A-206A; doi: 10.1016/j.amjhyper.2004.03.549

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stroke, heart failure renal failure and peripheral vascular disease from hypertension. At present, coronary heart disease is relatively uncommon, probably because Blacks in SSA have lower cholesterol levels and higher high-density lipoprotein cholesterol levels compared to the Caucasian population group in the Western world, the Asian and Caucasian population in SSA and in African Americans. Although there are good studies on the response and tolerability of antihypertensive drugs in SSA, there are no long-term morbidity and mortality data available.

Recommendations of the International Forum for Hypertension Control and Prevention in Africa (IFHA) have been documented. It should be emphasised that, while it is important to consider the science of medicine for the treatment of hypertension, particular consideration should be given to cost-effectiveness and affordability because many countries in SSA have severe resource restraints. In some of them, the health budget per capita does not exceed US\$10 per year and this is insufficient to address the needs posed by the double burden of NCD and infectious diseases, including AIDS.

Key Words: Hypertension, Sub-Saharan Africa, Cardiovascular Control Program

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THE RISK OF CORONARY HEART DISEASE, STROKE OR TOTAL CARDIOVASCULAR EVENTS IN HYPERTENSIVE PATIENTS WITH HYPERCHOLESTEROLEMIA IN JAPANESE POPULATION - SUB-ANALYSIS OF THE J-LIT STUDY, A LARGE-SCALE OBSERVATIONAL COHORT STUDY

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The Japan lipid intervention trial (J-LIT) was primarily conducted to clarify the relationship between serum lipid levels and incidence of cardiovascular event (CE) in Japanese patients with hypercholesterolemia (Circ J 2002; 66: 1087). Hypertension is one of the major risk factors for coronary heart disease (CHD) or stroke. We analyzed the results of primary prevention cohort in the J-LIT study, focusing on the relationship between blood pressure (BP) and the risk of CE, stroke or total CE in patients with hypercholesterolemia.

All patients (n=40,229) were treated with open-labeled simvastatin mainly at a dose of 5 mg/day for 6 years. According to the control level of serum total cholesterol (TC), patients were divided into two groups, less-controlled (L) (TC \geq 220 mg/dL, n=5,548(male), 13,421(female)) and well-controlled (W) (TC < 220 mg/dL, n=7,039(male), 14,213(female)) groups. We examined the effect of BP during the lipid-lowering therapy on the risk of CE and stroke comparing L with W group. The primary endpoint of the sub-analysis was stroke or CE such as acute myocardial infarction or sudden cardiac death, or total CE (CE+stroke).

The average age was 53.9 \pm 9.1 in male and 59.4 \pm 6.5 years old in female. The incidences of CE and stroke under lipid lowering therapy were 1.47 and 3.05 in male and 0.59 and 2.14 per 1,000 patients-years in female, respectively. The adjusted relative risk of total CE in the L group was increased at lower level of BP compared to the W group in both genders. The levels of systolic BP for total CE risk in the L group were \geq 130 mmHg in both genders. However those levels in the W group were \geq 140 mmHg in male and \geq 150 mmHg in female. The levels of diastolic BP for total CE risk in the L group were \geq 80 mmHg in both genders.

Those levels in the W group were \geq 90 mmHg in male and \geq 80 mmHg in female.

The risk of CE or stroke was increased with increase of BP in patients with hypercholesterolemia regardless the controlled level of TC. For patients with hypercholesterolemia and hypertension, we concluded that BP should be controlled strictly for the prevention of CE or stroke in addition to lowering serum TC.

Key Words: Cholesterol-lowering medication, Cardiovascular event, Blood Pressure

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UNCONTROLLED HYPERTENSION IN FABRY DISEASE

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Fabry disease is a x-linked lysosomal storage disease leading to early death related to renal, cardiac, and cerebrovascular disease. Therefore, proper diagnosis and therapy of elevated blood pressure may improve morbidity and mortality of these patients. However, the prevalence of uncontrolled hypertension in Fabry disease is unknown.

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Our study revealed a high prevalence of uncontrolled hypertension among patients with Fabry disease. Thus, there is need for improvement of blood pressure control in these patients.

Key Words: Fabry Disease, Uncontrolled Blood Pressure,